Mature cystic teratoma of suprahepatic localization, presentation of case and literature review

Teratoma quístico maduro de localización suprahepática. Presentación de caso y revisión de la literatura

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Abstract

We present the case of a suprahepatic tumor related to mature cystic teratoma, extragonadal germ cell tumors are rare and represent from 1.6 to 5% of all germ cell tumors, the most common site and the extragonadal presentation is the mediastinum (50-70%), retroperitoneum (30-40%), other rare locations pineal gland, sacrococcal region, prostate and bladder; however, the supra-hepatic location is not so common and is not documented in the literature.

Key words: Abdominal tumors. Extragonadal germ cell tumors. Mature cystic teratoma. Suprahepatic tumor.

Resumen

El tumor suprahepático se relaciona con el teratoma quístico maduro. Los tumores de células germinales extragonadales son raros y representan del 1,6 al 5% de todos los tumores de células germinales. El sitio más común y la presentación extragonadal es el mediastino (50-70%), seguido del retroperitoneo (30-40%) Otras localizaciones raras son la glándula pineal, la región sacrococcígea, la próstata y la vejiga; sin embargo, la localización suprahepática no es tan común y no está documentada en la literatura.

Palabras clave: Tumores abdominales. Tumores de células germinales extragonadales. Teratoma quístico maduro. Tumor suprahepático.
Case report

Clinical history and findings

This is a 39-year-old female who is referred from second-level hospital, due to liver tumor, for diagnosis and treatment.

The previously healthy patient presented to her reference hospital, due to the presence of a palpable mass since March 2017, in the upper right quadrant with progressive growth. With shooting pain since August 2017 increased in intensity and frequency, without irradiation, general malaise, fever of unknown origin without schedule, weight loss of 5 kg for 4 months, asthenia and adynamia. Treated in ixtlahuaca hospital without pain improvement for what is referred to this unit.

Pathological history: Surgical history resection of right ovarian cyst in 2016, with no known histopathological report. Without chronic degenerative diseases, she denies transfusions, alcoholism and smoking.

Gynecological history: menarche at 14 years old, pregnancies: 6; deliveries: 6; abortions: 0; Pap smear 1 year ago without malignancy results. Mammography 1 year ago without pathological data, 2016 oophorectomy secondary to right ovarian cyst.

- Admission laboratory studies November 8, 2017
  Glucose 99 mg/dl, bun 9 mg/dl, urea 19.26 mg/dl, creat 0.5 mg/dl, uric acid 4.4 mg/dl, cholesterol 113 mg/dl, proteins 6.5 g/dl, ggt 99u/l, ast 36 u/l, alt 36 u/l, fa 185 u/l, dhl 1219 u/l, amylase 30, lipase 28, sodium 141 mmol/l, potassium 4 mmol/l, chlorine 107 mmol/l, phosphorus 4 mg/dl, magnesium 2.1 mg/dl, calcium 8.4 mg/dl, total bilirubin 0.8 mg/dl, direct bilirubin 0.2 mg/dl, indirect bilirubin 0.6 mg/dl, leukocytes 7.6 x 10 (3), alb 3.4 g/dl 09-11-17 Ca 125 = 485 u/ml, Ca 19-9 = 25.16 u/ml, carcinoembryonic antigen 2.82 ng/ml, and alpha-fetoprotein 3622 ng/ml.

Image result

Tomography: tumor heterogeneous solid and cystic nature that compress the hepatic parenchyma, without invading it, in contact with the hepatic segments VIII, VII, and VI, gastric cavity deployed, as well as abdominal viscera, compression of the portal system (Fig. 1).

Intrahospital procedure

Due to the tomographic and biochemical findings, she was scheduled to laparotomy exploratory and...
tumor resection. Surgical procedure was performed with a midline incision type makuuchi, finding a tumor of 30 cm by 15 cm by 10 cm, in the inferior region of the right diaphragmatic pillar and invading hepatic segments VIII, VII, and VI, of a cystic aspect. Adhered to the liver without invading it, a complete resection of the tumor was performed, with an electrocautery, and foam gel was placed on the bed with hemostatic stitches (Figs. 2 y 3).

Histopathological report January 2, 2018

Microscopic description: germinal neoplasm constituted by cystic areas, lined by keratinized stratified squamous epithelium (epidermis) and respiratory epithelium with fragments of hyaline cartilage and cylindrical epithelium areas with mature adipose tissue, other segments constituted by cubic cells with eosinophilic cytoplasm and hyaline globules, in glandular pattern with Schiller-Duval bodies.

Final report: mixed germ cell tumor with mature cystic teratoma component (70%), endodermal sinus tumor (30%), and glandular pattern (Figs. 4 y 5).

Results and outpatient following

The patient remains hospitalized for 3 days, the oral route was started the day after surgery with adequate tolerance, and she was discharged without complications. At this moment after 1 year of follow-up, she does not present data on tumor relapse or imaging findings, liver function tests without alterations.

Discussion

Germ cell tumors prevail in females aged 10-30 years, accounting for 70% of ovarian neoplasms in this group. Our patient was 39 years old.

Extragonadal germ cell tumors are rare and represent from 1.6 to 5% of all germ cell tumors, the most common site of extragonadal presentation is the mediastinum (50-70%), retroperitoneum (30-40%), other rare locations pineal gland, sacrococcygeal region, prostate, and bladder; however, the supra-hepatic location is not so common and is not documented in the literature. Mature cystic teratoma accounts for 95% of all ovarian teratomas and they are mostly benign.
Most tumors are solid but may appear heterogeneous collections, may contain tissue and organized structures derived from the three cell layers, are mostly unilateral, and may be benign; however, there may be peritoneal implants4,5.

**Histopathology**

Mature cystic teratomas contain ectodermal, mesodermal and endodermal tissue, the mechanism of development is due to possible failures of meiosis II or the premeiotic cell in which meiosis I failed.

Macroscopically, it has the appearance of a multicystic mass that contains hair, teeth, skin, adipose tissue, and various tissues (lung tissue, and glandular tissue); in this case, the histopathological report is shown squamous epithelium (epidermis) and respiratory epithelium with fragments of hyaline cartilage and cylindrical epithelium areas with mature adipose tissue.

The diagnosis is made confirming the histology, through the presence of ectodermal, mesodermal, and/or endodermal tissue.

**Clinical manifestations**

Patients usually represent one or more of the following symptoms, abdominal growth (tumor), abdominal pain, transvaginal bleeding abnormalities, pregnancy symptoms, and 85% of patients have abdominal pain and abdominal mass. In case of ovarian tumor patients usually represent one or more of the following symptoms, abdominal growth (tumor), abdominal pain, transvaginal bleeding abnormalities, pregnancy symptoms, 85% of patients have both abdominal pain and abdominal mass.

In the case of extragonadal tumors, the manifestations are related to the increase in volume and compressive symptoms, whether mediastinal, retroperitoneal, intracranial, or abdominal. Our patient complains of abdominal pain, abdominal growth, general malaise, fever, weight loss, all of them unspecific symptoms, and preoperative diagnosis are often difficult, in case of abdominal presentation it may be consider as a differential diagnosis6,7.

**Laboratory**

Liver function and preoperative studies should be performed. Differentiated teratomas or mature does not produce increased α-fetoprotein (FP) or β-human chorionic gonadotrop, in this case, elevation of α-FP may respond to liver damage secondary to compression and not tumor activity, it was corroborated in the follow-up with the normalization of α-FP.

**Radiology**

Computed tomography scans are known to be more effective than ultrasound in diagnosing teratomas that occur in the mediastinum or retroperitoneum. Theoretically, distinct imaging features, such as the heterogeneous solid and cystic nature of the mass that sometimes have additional fatty or calcific elements, can help characterize and differentiate teratomas from those entities, depending on its size it can displace adjacent structures. These characteristics are similar to those found in our patient.

MRI typically provides harmless and favorable imaging delineation. The typical appearance is a well-circumscribed complex lesion with both solid and unilocular or multilocular cystic components8-10.

**Treatment**

The treatment of choice in all cases is surgical, since the mature benign tissue does not responds to chemotherapy, the key is to remove all the lesion while preserving the normal structures, if necessary at least margins of 0.5 cm. In addition, delay in treatment may result in interim lesion growth, infection, or malignant transformation. An extensive surgical approach should be performed to avoid persistence of the tumor at the surgical site and achieve R0 resection.

Among the postsurgical complications, bile leak, bleeding, and lymphatic injury are the most common9-11.

**Follow-up**

The recurrences of benign teratomas following complete surgical resection are rare, and the long-term prognosis is excellent.

Malignant transformation occurs in 0.2-2% of mature cystic teratomas, comprise 2.9% of all ovarian tumors dependent on germ cells, even though any component of the tumor can develop a malignant disease, most often is that the squamous cell, be the one that has this transformation.

Risk factors for malignancy are: patient > 45 years old, tumor diameter > 10 cm, rapid growth, imaging findings12-15.
Our patient after 1 year of follow-up, she does not present data on tumor relapse or imaging findings, liver function tests without alterations.

**Conclusion**

The present case involves only one patient, and because there are very few case reports, it is not clear whether the discussion of the present case can be applied to all cases. In some cases, the appearance of a tumor that was implanted in the liver secondary to a primary ovarian tumor is explained by the hepatic vascularization and by the drainage pattern of the peritoneal fluid.

Although it is not a frequent pathology, it represents a differential diagnosis that should be taken into account in young women.

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**Conflicts of interest**

The authors declare that they have no their conflicts of interest.

**Ethical disclosures**

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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